

Hemiplegia in an 11-Year-Old

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Patient Report

An 11-year-old right-handed, otherwise healthy girl was admitted to a community hospital for right upper and lower extremity weakness of sudden onset. Three days before she was presented to the hospital, she complained of headache, rated as moderate to severe, described as pressure over the bifrontal area, with no radiation, and not associated with photophobia or phonophobia. There was no accompanying nausea or vomiting. The pain was relieved by acetaminophen briefly, but the headache persisted intermittently until the day she was seen in the emergency department. When asked, she said she did not have similar headaches in the past.

On the day of the event, the patient came home from school and played a video game (*Dance Dance Revolution*, a music video game series produced by Konami Digital Entertainment Co., Ltd, Tokyo, Japan) with her classmate, after which she walked over to the couch to rest. The mother was called a few minutes later by her friend who noted that the patient had rolled over and fallen on the ground. When her mother saw her she was staring blankly with no verbal output. It was also noted that she was not moving her right side. She was thereafter transported to a community hospital.

On examination she had vitiligo on her skin. Good carotid flow was present with no bruits. Pulses were normally palpable in all the 4 extremities. Cardiac auscultation revealed a regular rate and

rhythm. On neurologic examination, she was awake and alert. She had expressive aphasia but was able to utter some words including "fine" and "good" when asked how she was doing. Cranial nerve examination revealed a decrease in facial sensation on the right side in the distribution of all 3 branches of the trigeminal nerve and an upper motor neuron pattern facial weakness on the right. Motor examination revealed a dense right hemiplegia. Reflexes were brisker on the right three-fourths compared with the left, with an equivocal plantar response on the right. Sensory examination showed a decreased sensation on the right side with respect to all primary modalities and an impairment of graphesthesia and a two-point tactile discrimination.

Computed tomographic scanning of the brain and magnetic resonance imaging (MRI) were done within 1 hour of admission, and the results were found to be normal. She was given a diagnosis of hemiplegic migraine. The following day, as the motor weakness persisted, she was transferred to our institution. A repeat MRI done 3 days later showed an infarct in the territory of the left middle cerebral artery and concomitant magnetic resonance angiography showed a "double lumen" within the left internal carotid artery (Figures 1A and 1B). An angiogram done subsequently confirmed the lesion.

She recovered to strength of 4/5 (movement against gravity and some resistance) in terms of motor weakness with residual facial weakness on a 6-week follow-up visit. She was independent with all activities of day-to-day living. Speech also recovered to a large extent with ability to participate in conversations but with occasional word-finding difficulty. At no point did she have visual symptoms indicative of ophthalmic artery involvement or did she have lesions in the territory of the anterior cerebral artery.

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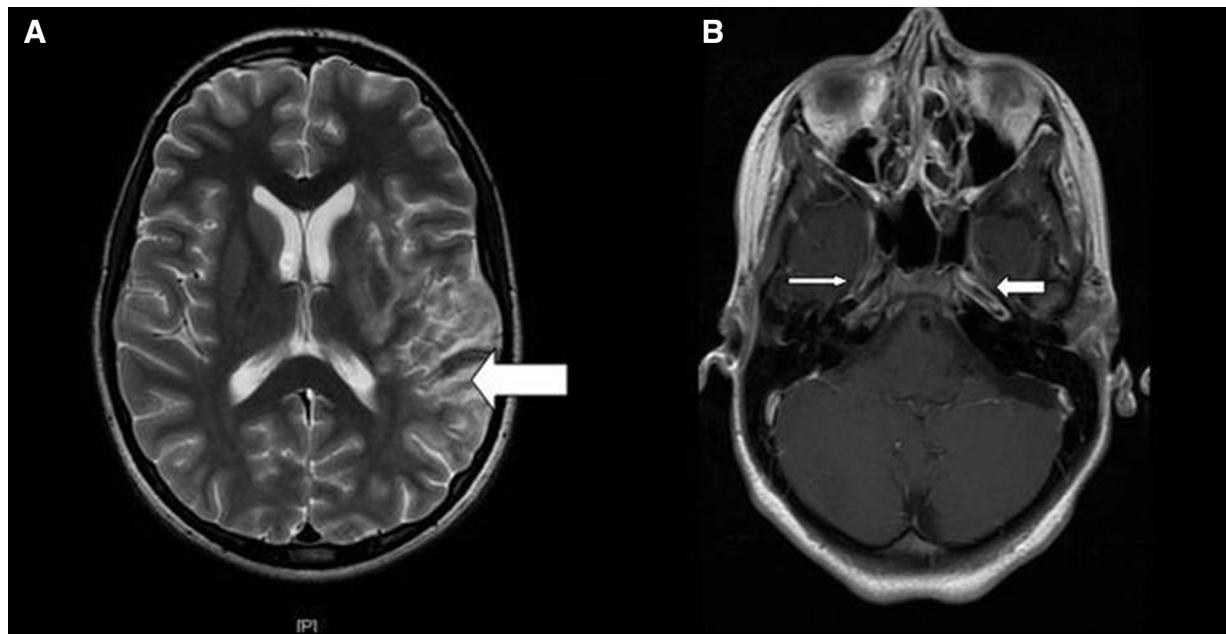


Figure 1. A, Brain magnetic resonance imaging. Axial fluid attenuated inversion recovery (FLAIR) image showing lesion in the left middle cerebral artery territory. Arrow indicates bright lesion in the parenchyma suggestive of underlying infarction. B, Brain magnetic resonance angiography. Bold arrow indicates “double lumen” within the left internal carotid artery.

Diagnosis: Internal Carotid Artery Dissection

Discussion

Carotid artery dissection is considered a rare occurrence in childhood, though in certain series the reported incidence among those children presenting with a stroke is as high as 7.5%.¹ The incidence of childhood stroke in the United States is believed to be 2.5 to 3/100 000, with case mortality rates ranging from 7% to 28%.² Because a clear-cut etiology is identifiable in less than half the cases, recognizing dissections is of importance in terms of planning treatment.

Extracranial dissection is far more frequent because of the anatomic proclivity of cervical vessels to injury. A review of 118 cases of dissection in childhood, over a period ranging from 1964 to 2000, indicates that most of the anterior circulation dissections are intracranial when not preceded by significant trauma.³

Dissections usually involve an intimal tear, with blood entering the vessel wall under pressure, leading to occlusion of the lumen and the resultant

ischemia. This leads to the characteristic features on imaging of an intimal pouch. Less frequently, dissections may entail a tear in the media with entry of blood between adventitia and media. This can lead to aneurysmal dilatation or vascular rupture. When the intracranial segment of the carotid artery is involved, stenosis usually stops short of the petrous segment.⁴

Interestingly, a male preponderance was noted, even when trauma or physical activity was not implicated. In this case, although she was taking part in a vigorous physical activity, no direct trauma could be identified which unfortunately lead to a lower index of suspicion for this condition. A minor precipitating factor is not unusual in case reports where instances of painting a ceiling, vomiting, sneezing, or unusual postures is reported during yoga preceding an event. Occurrence during chiropractic manipulation has been widely recognized as a predisposing factor for cervical dissection.⁵ In most instances, clear-cut documentation of cause and effect is lacking as most studies are retrospective in nature, with inherent recollection bias.

The presence of conditions which may be associated with structural defects of the vessel wall such as Ehlers-Danlos or Marfan disease increases the

chances of spontaneous events. Ehlers-Danlos type IV is characterized by abnormalities in the structure of collagen and the increase in mortality attributed to rupture of splenic or abdominal vessels. Arterial aneurysms can be a comorbidity. In fact, in the case series of Ulbricht et al,⁶ skin biopsies from 6 out of 7 patients, with spontaneous carotid artery dissections, showed abnormalities in the connective tissue matrix viewed under electron microscope, comprising twisted collagen fibrils, with variation in diameter and disorganization. Fibromuscular dysplasia has been reported in up to 15% of adult patients with dissections, though similar literature for children is lacking. Other coexistent conditions include osteogenesis imperfecta, bicuspid aortic valve, and aortic coarctation. Preceding upper respiratory infections have been implicated as well.⁷

The clinical features in almost all reported cases include a focal deficit secondary to ischemia and headache in about 50% of cases. The headache preceded other symptoms by hours to days.⁸ The headache has variably been described as migraineous, cluster-like, mimicking hemicrania continua or unilateral neuralgiform. Headaches are an unusual feature of strokes caused by other conditions and hence should serve as a red flag for hemorrhagic strokes and dissections. Subarachnoid hemorrhage may be a rare presenting manifestation.⁸

Confirmatory diagnosis was heavily dependent on direct visualization by catheter angiography though now MRI can be relied on to provide similar data.^{9,10} The essential complications of dissections on angiogram is the demonstration of an intimal flap and/or a mural hematoma though subsequent complications may include stenosis or formation of an aneurysm.

About 17% of patients, a relative minority, have a history of physical activity preceding the event, which could range from playing soccer to dancing. The average time lag between the trauma or activity and the development of neurological deficits varied from 1 to 7 days in most instances, though a very brief interval of a few minutes to immediate has also been rarely described.

The optimal treatment of this condition also remains obscure due to the relative scarcity of the literature, though the case series of Fullerton et al³ does indicate a higher mortality in those not treated with anticoagulant. Pharmacological management includes treatment with anticoagulants,

most popularly warfarin, and antiplatelet agents. The use of low molecular weight heparin has not been adequately evaluated. In most instances, this treatment pertains to cervical dissections as intracranial extension does constitute a relative contraindication to anticoagulation. When anticoagulation is instated it is usually continued for 3 to 6 months, with high recanalization rates observed in 2 to 3 months. Another approach is to use antiplatelet therapy alone, especially when ischemic symptoms are absent. Again, the duration of treatment remains a matter of conjecture and debate. Use of anticoagulants does not alter the outcome with respect to recurrence.

Recently, there has been interest in using endovascular techniques such as percutaneous angioplasty and stenting, though its use in children remains to this date experimental.¹¹ Furthermore, stenting has been an approach used almost exclusively in cervical carotid artery dissections. Low complication rates appear to make this a viable option in certain scenarios.

Finally, surgical techniques such as arterial ligations and grafting of the affected segment have been attempted.

The literature is unfortunately sparse with respect to this condition which can carry a high mortality rate of up to 50%.³ Children with intracranial involvement have a poorer outcome than those with cervical pathology. The largest case series of spontaneous intracranial dissection has 18 patients³ and provides an excellent review regarding this very important neurological condition as does the report by Schievink et al.⁴

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